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Treatment of a localized exogastric gastrointestinal stromal tumour (GIST) with long-term follow up: A case report and review of literature

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Abstract

Background: Gastrointestinal stromal tumour (GIST) is a rare entity. The diagnosis, treatment and follow-up pose a challenge in our environment. We describe the presentation, diagnosis, treatment, and long-term follow-up of a patient with a localized, large exogastric GIST.
Method: The case notes of a 51-year-old woman who presented with a one-year history of upper abdominal swelling with no associated symptom was reviewed.
Result: On examination, an oval intraperitoneal mass at the epigastrium measuring 16 x 14cm which was freely mobile, smooth, firm, non-tender with dull percussion notes was found. She subsequently had resection of a large exogastric mass with a histologic diagnosis of GIST. Following tumour resection with a wide margin of gastric wall, she has done well so far for ten (10) years.
Conclusion: Wide margin gastric resection may suffice in the treatment of a large localized gastric GIST when it is exogastric.

Keywords: GIST, gastric, exogastric, large mass

Introduction

Gastrointestinal stromal tumour (GIST) represents only 1% of all malignant tumours of the gastrointestinal tract (GIT).[1] It is, however, the most common mesenchymal tumour of the GIT with majority (47-60%) arising from the stomach and small intestine and a small number in other parts of the GIT.[2] Gastric GIST originating from the wall of the stomach may present diagnostic and therapeutic challenges. This report describes the case of a patient with a localized gastric GIST treated only by surgical resection followed by a decade of monitoring.

Case report

A 51-year-old woman presented with a one-year history of upper abdominal swelling. There was no history of easy satiety, vomiting, haematemesis, malaena stool or weight loss. She was not pale and there was no peripheral lymphadenopathy. Abdominal examination revealed an oval intraperitoneal mass measuring about 16 x 14cm at the epigastrium; firm, smooth, freely mobile, not tender, and dull on percussion. There was no demonstrable ascites. Abdominopelvic ultrasound scan revealed a well circumscribed oval mass in the epigastric region anterior to the inferior vena cava and medial to the gall bladder, bounded laterally by the stomach. The mass measured 10.2x11.8cm with mixed echogenicity and a central area of hypoechogenicity (representing area of necrosis). The liver had normal parenchymal echogenicity with no space occupying lesion noted. She was unable to do computerized tomography scan as the machines in Umuahia and Port-Harcourt were faulty.

She underwent laparotomy. Intraoperative finding was a pedunculated, spherical gastric tumor,

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firm/hard with vascular markings (figure 1) measuring 13x10x10cm weighing about 100g. She underwent resection of the gastric tumour with a margin of normal gastric tissue, and gastric repair was done.

Postoperatively, she was commenced on graded oral intake on the fourth day. She was discharged home ten days after surgery to be seen on out-patient basis. Histology of the specimen showed sheets of spindle-shaped cells arranged in randomly oriented fascicles.

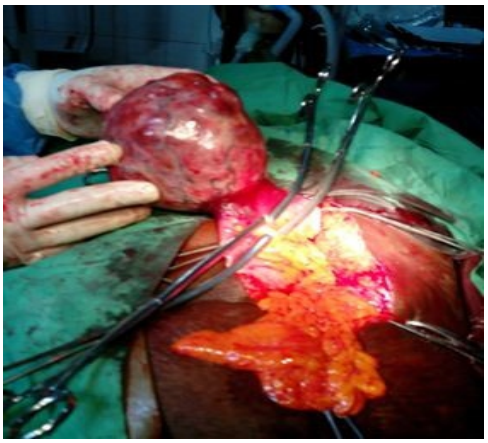
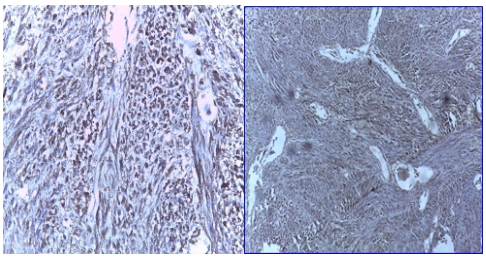


Figure 1: Intraoperative picture showing a localized gastric mass



Figures 2A and 2B: Showing the immunohistochemistry slides

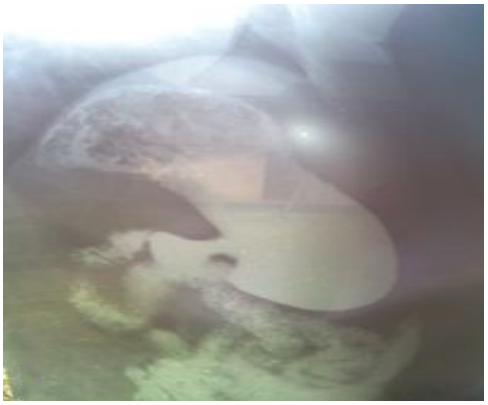


Figure 3: Barium meal and follow through done 12 months after surgical resection of the mass

The cells were pleomorphic and exhibited hyperchromasia interspersed with occasional mitosis. The diagnosis was GIST (Gastric tumour).

The immunohistochemistry using Ventana antibody with in-house positive control is as follows: C-KIT – moderately reactive (figures 2A and 2B); CD 34 – weakly reactive; S100 – Negative; CD 45 – Negative; EMA – Negative. Barium meal and follow through done 12 months after surgical resection revealed no abnormality (figure 3). She has remained symptom-free for ten (10) years with no evidence of recurrence.

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Discussion

Gastrointestinal stromal tumours (GISTs) are non-epithelial, mesenchymal tumours. GISTs were first described by Mazur and Clark in 1983 and are thought to arise from interstitial cells of Cajal (ICC) or their stem cell precursors which are normally part of the autonomic nervous system of the intestine.[2] They are most common in the stomach (60-70%), followed by small intestine (20-25%), colon and rectum(5%),and oesophagus (<5%).[3,6] GIST can develop outside the intestinal tract, within the abdominopelvic cavity such as the omentum, mesentery, uterus, vagina, and the retroperitoneum; they are called extragastrointestinal stromal tumours (eGIST), usually behaving aggressively.[2,4] Benign tumours outnumber the malignant ones by a wide margin. 10-30% are said to be clinically malignant.[4] Borderline GIST has been described; morphologically, they typically exhibit moderate cellularity, and subsets of them show moderate atypia, low mitotic activities or large tumour size.[5,6] Our patient had the borderline variety. Peak age of occurrence is 60 years.[1] It has been shown to affect more men (55%) than women.[2] GIST may present in different ways depending largely on the size and location of the lesion. Seventy percent of patients have symptoms. The others are diagnosed incidentally or at autopsy. Symptoms may include gastrointestinal bleeding (acutely, as in melena; haematemesis, or chronic insidious bleeding); mass effect may present as easy satiety, bloating and abdominal pain.[2,7] Others may include nausea,

pleuritic chest pain, bowel obstruction, altered bowel habit and weight loss. Metastatic sites are commonly the abdominal cavity and liver, rarely bones and soft tissues and very rarely lymph nodes and skin.[2,7,8] Our patient only complained of upper abdominal swelling and there was no evidence of infilteration of adjacent structures or metastasis. GIST are associated with some tumour syndromes in less than 5% of cases; these are familial GIST, neurofibromatosis type 1, Carney's triad and Carney-Stratakis triad.[2,9,13] Currently, GIST can be routinely identified using histological, immunohistochemical, and molecular genetic assays.[10,11] Contrast-enhanced computerized tomography is the standard method for GIST imaging.[12] We could not do CT as the machines in Umuahia and Port-Harcourt were faulty. Magnetic resonance imaging is indicated in liver-specific questions or CT is contraindicated. Chest imaging, endoscopic ultrasound scan, and endoscopy are also done.[1] PET/CT scan is used for early response assessment, density and appearance of new lesion.[2,8] We did not biopsy the mass as this could lead to rupture which increases risk, and likelihood of recurrence. They also show a wide clinico-pathological spectrum, from a small probably incidental nodule to a large pedunculated mass which our patient had. They are usually tan to white, well circumscribed lesions. Histological cell types include spindle cell type which was also noted in our study account for 70% of cases; others are the epithelioid cell type, and the mixed cell type.[4,13] Greater than 95% of GISTS are positive for CD117/KIT other less specific antigens CD 34, nestin, caldesmon, calponin, and vimetin.[13,14] Ki-67 expression in GISTs have significantly been associated with the size of the tumours, mitotic rate, and risk of malignancy.[15] Tumour size, location, mitotic index and tumour rupture have been used in risk stratification.[8,16,17] Any GIST >10cm qualifies for high risk according to the National Institute of Health consensus criteria. In a study done which considered large GISTs, the median size and mitotic count were 12.5cm, 8/50HPF.[18] Surgical resection with the healthy border of stomach tissue may suffice as primary treatment in a localized gastric GIST.[19,20] In our patient, the tumour was large but pedunculated and extraluminal (exogastric). We resected the localized gastric GIST with 5cm of normal stomach tissue, and did a gastric repair. Surgical resection may also be done after neo-adjuvant therapy to shrink tumour size; debulking surgery may be done for symptomatic relief. They respond poorly to conventional chemotherapy and radiation therapy.[21,22] Imatinib mesylate and Sunitib maleate competitively inhibit KIT and PDGFRA.[23] Imatinib has been shown to induce an extended remission in patients with advanced disease, most of whom would have been dead Within a few months of diagnosis.[24,25] Sunitib is a second line therapy for advanced GIST after Imatinib resistance and/or tolerance.[2] Gastric GIST have a lower risk of recurrence than non-gastric tumours of the same size and mitotic count.[26] Some workers noted a median relapse free survival time after surgery as fifty months, however, tumour mitotic count >5/50HPF, size >5cm, non-gastric location, tumour rupture, and male gender have adverse influence on relapse free survival.[27] We have followed up our patient for 10 years without relapse. Long term follow up is important due to the potential for late recurrences.[28]

Conclusion

Surgical resection with the healthy border of stomach tissue as primary treatment of localized GIST is recognized.[19,20] Limited gastric resection with tumour-free margins may suffice in the treatment of a large localized gastric GIST when it is exogastric.

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